Title: **A Comparative study: feature and clinical progress of Familial Mediterranean Fever to clinical manifestation onset in 1st decade of life and upper decades**

ABSTRACT

BACKGROUND: Familial Mediterranean Fever (FMF) is an autosomal recessive disease characterized by recurrent episode of serositis attack, commonly involving the abdomen, chest or joint, typically accompanied by fever and elevated acute phase reactants. Attacks subside spontaneously within two to three day without residue.

The disease affect people living around Mediterranean sea (i.e. Turks , Armenians , Arabs and Jews) and cause by a number of mutation in MEFV gene on the short arm of chromosome 16, coding for the protein "pyrin". Continuous treatments with colchicine reduce attacks frequency, duration and prevent the development of secondary Amyloidosis. In this study, we compare the manifestation and clinical progress when FMF appearance in 1st decade of life and upper decades.

METHODS: Two hundred twenty patients referred to the clinic of FMF in Boo Ali hospital of Ardebil. A clinical diagnosis of FMF was made according to published criteria, patients divided to two groups at sign appearance in 1st decade of life and upper decades. Compare the demographic, clinical characteristics, response to colchicine therapy and side effect of colchicine between these groups.

RESULTS: 220 patients participate in this study those 144 patients (65.5%) of them sign appearance in 1st decade of life and male to female ratio in first group is 1.21 and in second group is 1.29 and average of period of pain and duration of pain in first group respectively is 30.70 days and 2.28 days and in second group is 40.04 days and 2.70 days.

Fever (95.8% vs. 86.8 % : p= 0.015) , Abdominal pain (91% vs. 90.8% : p=0.964) , Anorexia (72.9% vs. 86.8% : p=0.018) , Myalgia (18.1% vs. 30.3% : p= 0/039) and Arthritis (12.5% vs. 30.3% : p=0.001) . Good response to colchicine therapy in first group is 83.33% and in second group is 86.84%.

Altogether 45 patients from 61 patients that gene analysis homozygote and heterozygote (two mutations) and most common mutation are M694V and then M680I.

CONCLUSIONS: Duration of pain and fever are more than and anorexia , myalgia and arthritis is less than when sign appearance in 1st decade of life and M680I mutation is most common in 1st decade.

Key words: Familial Mediterranean Fever, clinical progress, MEFV gene